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The impact of cystic fibrosis on recovery from orthopedic trauma: A case study

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Abstract

Cystic Fibrosis has effects on many body systems, including the skeletal system. In this case, we describe the impact of respiratory and endocrine disease on bone healing following orthopedic trauma in a 22-year-old woman. Limitations to mobility resulting from trauma complicated her respiratory condition. Disease-related effects on bone health and healing delayed her recovery from the trauma. It is important to recognize the multisystemic nature of CF, even when managing acute orthopedic injuries.

Keywords: Cystic fibrosis; Fractures; Bone

1. Introduction

Cystic Fibrosis (CF) has historically been considered primarily a disease of the lungs and pancreas. Recently, however, there is evidence that many systems are affected by this disease, including the musculoskeletal system [1,2]. Patients with CF have been found to have decreased bone density, increased risk of osteoporotic fractures and other bone and joint effects [2,3]. These findings suggest that the disease affects bone mass gain and maintenance. In addition, injuries of the chest wall and proximal upper extremities could impact the respiratory status of these patients, though interaction with breathing and airway clearance techniques. In this case report, we discuss the management of a young woman with CF who was hit by an automobile, causing multiple fractures and how the management of her recovery from this orthopedic trauma was affected by the manifestations of CF.

2. Case report

CL was a 22-year-old young woman who was hit by a car while crossing the street. She incurred fractures of the right scapula (Fig. 1), right tibia and fibula, left medial malleolus and left fifth finger. She was admitted from the emergency department to the hospital. She underwent surgical fixation of the lower extremity fractures only. Her baseline CF status involved severe lung disease, with a forced expiratory volume in 1 second (FEV_1) of 31% of predicted, multiple drug-resistant lung organisms, and CF-related diabetes. Seven months prior to the injury, she had a bone density (DEXA) scan that revealed a *T*-score for the lumbar spine of − 3.94. In addition to the usual supplemental fat-soluble vitamins she had taken for many years (800 units Vitamin D twice daily), she was prescribed alendronate 10 mg daily, vitamin D 800 units daily and calcium 1500 mg daily. Lab values for serum vitamin D levels were unavailable due to an oversight.

Her hospital course was complicated by difficulties performing airway clearance, due to scapular fracture pain and respiratory suppression from narcotic medications. The patient developed respiratory failure, with pCO_2 values above 60 mm Hg, and required nasal positive airway pressure support for several days. She was also treated with an

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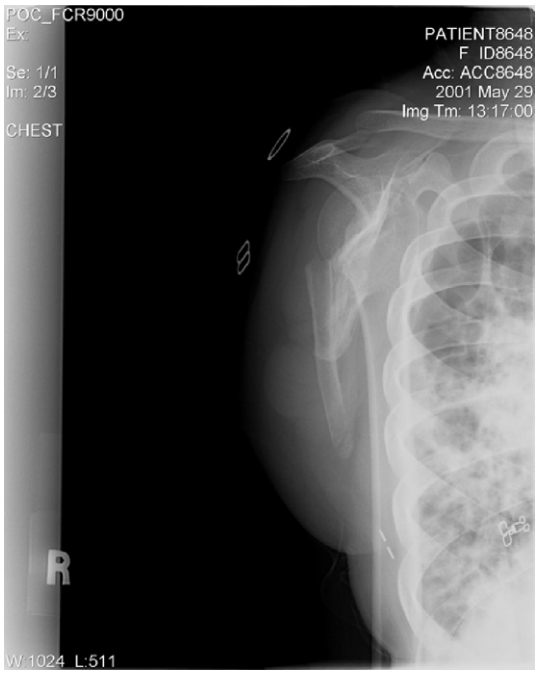

Fig. 1. Radiograph showing scapular fracture as well as extensive disease in the right lung.

extended course of intravenous antibiotics. Her pulmonary status was further compromised by the inactivity imposed by non-weight bearing status on both lower extremities. Thus, her mobility was limited to being lifted from bed to chair. After 40 days of hospital care, she was discharged home, confined to a wheelchair.

Adequate pain control was very difficult to achieve in this patient. She required high levels of narcotics in order to manage her pain, up to two years following the accident. She then developed a dependence on these medications and exhibited drug-seeking behaviors. The use of narcotics was likely a factor in her frequent episodes of constipation.

Fracture healing was slow, and weight-bearing status reflected this. She was finally allowed to begin weight-bearing on her lower extremities four months post-fracture, however, delayed fracture healing was evident on X-ray three months later (Fig. 2). Her healing was also complicated by a broken screw, which was eventually removed. Her scapular fracture impaired her left arm motion to such a degree that she developed frozen shoulder syndrome. Physical therapy was initiated to improve arm mobility and progress ambulation as well as exercises in the pool. The patient tolerated these programs fairly well and progressed slowly.

Two years following the accident, the patient was fully ambulatory and had regained full shoulder motion. However, her lung disease continued to progress. She is currently dependent on supplemental oxygen and awaiting lung transplantation.

3. Discussion

This case illustrates the interdependence of pulmonary and musculoskeletal manifestations of CF. We clearly observed the importance of physical mobility in the maintenance of lung function for this patient. When she was immobilized by her trauma, she quickly developed respiratory decompensation. In addition, intolerance to her preferred methods of airway clearance (vest, percussion) limited options for clearing secretions from her lungs. All of these factors, added to the effects of pain and narcotics, could easily have led to her death.

There were also long-term issues related to bone healing that were affected by CF. Pancreatic insufficiency common in CF impairs uptake of Vitamin D as well as absorption of calcium from fatty dairy food sources [1,4]. In this patient, it was found that normal supplements were not sufficient to prevent osteoporosis. Despite the addition of increased vitamin D, calcium and alendronate, her bone density was still poor at the time of the accident. These deficiencies can impair bone deposition. Inflammatory cytokines, released into the circulation in patients with lung exacerbations and resulting from insulin resistance can stimulate osteoclasts and accelerate bone resorption [5]. Decreased bone density (*T* scores less than −1.0 SD) has been seen in up to 85% of adults with CF [6]. These factors, added to the effects of immobility, may have explained the delayed healing seen in our patient's fractures.

Bone pain is found to occur in 2–8.5% of patients with CF [2]. Although our patient did not report bone pain prior

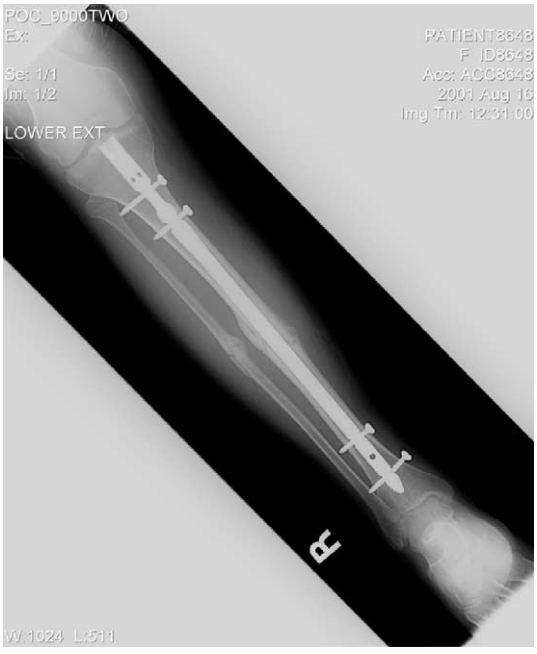

Fig. 2. Radiograph of tibia/fibular fractures 7 months post injury. Fractures are still clearly visible.

to the trauma, it is possible that she experienced greater bone pain than would be seen in a young woman without CF. The needs for high levels of narcotics unfortunately led to drug addiction, which further prolonged her recovery.

4. Conclusion

Through our experience with this patient, we learned several important points. First, CF is a multisystemic disease, which can impact both acute and long-term management of traumatic injuries. Patients with severe lung disease are at great risk for developing complications of immobility. A team effort is essential in managing the orthopedic, digestive, respiratory and endocrine effects of trauma in patients with CF and providing for the optimal functional recovery. Finally, it is vital that patients be familiar with multiple methods of airway clearance in case one method cannot be used.

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